Arresting pulmonary atresia intact ventricular septum...only if you have to

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Pulmonary atresia intact ventricular septum (PA-IVS) is a rare congenital cardiac anomaly composed of varying degrees of right ventricle (RV) and tricuspid valve hypoplasia. Perhaps most important, there are a wide range of coronary artery anomalies, including RV to coronary artery fistulas, coronary artery stenosis, and coronary artery atresia. RV to coronary artery fistulas are common, occurring in up to 75% of patients in some series.^{1,2} Coronary artery atresia, interruption, or severe stenosis may result in an RV-dependent coronary circulation (RVDCC) in which right and left myocardium are reliant on blood flow via fistulas from the RV. This coronary anatomy is rare, occurring in approximately 5% to 10% of patients.³ The literature is clear that patients with RVDCC have a poor prognosis, and consideration for primary transplantation should be given.

In this issue of the *Journal*, Dr Woods⁴ presents a high-risk patient with PA-IVS and RVDCC who was listed for cardiac transplantation after successful resuscitation from cardiac arrest while undergoing coronary angiography. Without a suitable donor and evidence of clinical deterioration, the infant was taken to the operating room for a successful atrial septectomy and bidirectional cavopulmonary shunt with pulmonary artery patch reconstruction. This report is important in that it offers an alternate technique description for successful cardioplegia administration in a patient with RVDCC. By simultaneously administering cardioplegia into the aorta and RV by way of the tricuspid valve, good coronary perfusion and cardiac arrest are obtained. It is reasonable to conclude that this method ensured cardioplegia penetrance into the postatrial or stenotic coronary regions that would have gone unprotected with simple antegrade administration.

Although this case report provides a potentially useful technique when faced with inducing arrest in a patient with PA-IVS and RVDCC, it should be remembered that typically we strive to avoid cardioplegic arrest in these patients. Catheter-based atrial septostomy may allow for staged palliation without cardioplegic arrest. While the

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Central Message

Delineation of coronary anatomy and avoiding arrest are important in PA-IVS. If arrest is necessary, dual aortic and right ventricular cardioplegia instillation offers a viable technique.

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patient is on bypass, keeping the RV full ensures adequate coronary perfusion. If arrest is required, one must remember coronary anatomy is highly inconsistent, and simple antegrade cardioplegia administration may result in inadequate myocardial protection. Other useful maneuvers include cooling on bypass and topical cooling of the heart with ice. Regardless of these techniques, an arrested heart with RVDCC may still struggle to regain the RV systolic pressure wave required to support coronary perfusion coming out of an arrested state.

Coronary anatomy in PA-IVS is highly variable, and it is critical that the anatomy is defined. In general, especially with the high-risk patients with RVDCC, cardioplegic arrest should be avoided if at all possible. If arrest is necessary, dual aortic and right ventricular cardioplegia instillation offers a viable technique.

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